Duhring (L. A.)

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A CASE OF THE SO-CALLED XERODERMA (OR PARCHMENT SKIN) OF HEBRA.[1]

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The disease of which the following case is an example is of such rare occurrence that any information respecting it must prove interesting and valuable. But very few cases have been recorded, and these quite lately, so that until a recent date no account of the disease whatever existed. The case before us differs in several points from those which have been described by Hebra and Kaposi and by Taylor, to which I shall refer later, and shows that the affection is not necessarily accompanied by such marked symptoms of atrophy as occurred in the two cases related by Hebra and Kaposi.

The patient, Annie McC., is a young woman, aged seventeen, of Irish parentage. Her father is living and is healthy; her mother is deceased, and is said to have died of cancer of the womb. She has three brothers and one sister living, and had four other sisters, who died quite young from various infantile diseases. Neither parent, nor any of the brothers or sisters exhibited any disease of the skin. There is no history of consumption.

According to the statement of the parents, the affection made its appearance at the age of six months, upon the face, more particularly over the nose and cheeks, in the form of ordinary freckles. At birth the skin was normal, and the child was considered healthy. The lesions gradually spread over the rest of the face, neck, and extremities, increasing in number from year to year. The early history of the disease cannot be ascertained more minutely, but it is said that at the age of nine years the lesions were quite as extensively distributed and as well-marked as at present, and that but little change has taken place since this date, except on the face and on the backs of the hands. Three years ago, when working in the mill and using some strong-smelling, offensive oil for lubricating the machinery, there appeared on the upper part of the cheeks about a dozen pin-head sized, solid, "reddish pimples." They came in the course of a week, and were confined to the cheeks. Some eight girls were similarly occupied with the machinery, all of whom were affected in the same man-

[1] Read at the Second Annual Meeting of the American Dermatological Association, at Saratoga Springs, Aug. 1878.

ner. The eruption disappeared within three or four weeks by scaling and slight crusting, and was succeeded in our patient, according to her statement, by the pock-like marks which still remain; but as far as she knows none of the other mill-hands were thus marked. She seems positive that the cheeks and nose were badly freckled before this eruption appeared, and that the atrophic marks followed, obliterating in a great measure the former freckles. Within the past five years she has noticed that on the backs of the hands the freckles have been here and there very slowly and gradually disappearing, and that in their place small areas of thinned whitish skin have come. No more particular information concerning the lesions can be obtained.

Present Condition.—The patient is tall and spare. She has dark-brown hair and eyes, and a fair skin upon the unaffected regions. The general nutrition of the body is good, the trunk and extremities being supplied with the average amount of adipose tissue. The general health is fair, and the various functions are normal. The sudoriparous system acts freely, but the sebiparous glands are somewhat sluggish in their action. The hair and nails are healthy, likewise the tongue and mucous membrane.

The disease occupies the scalp, face, ears, neck, chest, back, shoulders, arms and forearms, backs of hands and fingers, legs and backs of feet. It consists of an extensive, well-marked, disseminated, deposition of pigment, in the form of pin-head to split-pea sized, yellowish, brownish, and blackish, freckle-like lesions, together with the occurrence here and there of minute linear or rounded, pin-head sized telangiectases, and pin-head and split-pea sized, whitish, more or less defined, atrophic spots. There exist therefore three kinds of lesions to be considered, namely, pigmentary, telangiectasic, and atrophic.

The pigmentary deposit is very decidedly the most prominent feature of the disease, and is that for which the patient sought medical advice. All of the affected regions are thickly covered with what appear at first sight to be unusually defined, large and small, light and dark freckles, giving the skin a variegated, checkered look. The patient and her family believe the disease to be one of a singular form of freckles, and certainly the appearance is such that no other view would be entertained by any one were it not for the presence of the other lesions, to be presently described. The lentigines, or freckles, as I shall designate these lesions in speaking of them, exist in profusion, scarcely a portion of the affected regions escaping. They are disseminated, showing no regularity of distribution, nor any disposition to group. They are scattered over the surface at haphazard, as in the case of ordinary freckles. They are, however, as will be presently described, more abundant and more marked in some localities than in others. In size they vary from a small pin-head to a large split-pea, the majority perhaps averaging the size of large pin-heads. They are discrete or confluent, and are for the most part sharply defined, with very irregular, angular, jagged outlines, which cause them to stand forth conspicuously against the surrounding healthy skin. Their colour varies from light yellow to black; while the majority are of a dirty-yellowish, brownish-yellow colour, others are deep brown and some are quite black. The blackish lesions are, however, exceptional. The pigment is distributed unevenly, as in the case of ordinary lentigo.

Concerning the telangiectases, which, it may be here stated, are by no means numerous, it is observed that they occur here and there, and that they exist for the most part as small, ill-defined lesions. They are true

telangiectases, differing in no way from those encountered independent of other disease. They are made up of a congeries of capillaries. They are circumscribed or diffused, flat, raised, or slightly rounded, irregularly rounded, angular or linear, pin-head and even smaller sized, bright or deep red lesions. They are met with here and there over the whole of the invaded surface, but are most distinctly marked about the neck and chest. They are nowhere so numerous or so large as to be conspicuous, and they might readily be overlooked by the casual observer, or be regarded simply as accidental telangiectases. Occasionally only are they elevated, and seldom so to any extent, the majority being on a level with the surrounding skin.

The atrophic spots, like the telangiectases, are few and scattered, occurring here and there in between the freckles. They consist of pin-head and split-pea sized, rounded or irregularly shaped, somewhat glistening, whitish lesions. They bear resemblance to the marks of varicella or of variola, but they are more irregular in outline, more superficial, and less circumscribed and sharply defined. Taken between the fingers the skin is noted to be slightly thinned, to be soft and supple, and to have a smooth, polished surface, like glazed paper. The skin can be readily pinched up. Some of the lesions can be detected in passing the hand over them; in other places they are barely or not at all perceptible to the touch. Their shape and size in many instances are those of their neighbouring freckles. Here and there, as on the backs of the hands, there exist large, split-pea sized, irregularly shaped patches of atrophied skin, the result of several adjoining lesions having undergone simultaneous atrophy. The atrophy of the skin, however, viewing the process of atrophy as a whole, is insignificant, and is in some places so slight as to be scarcely appreciable.

The sensibility of the skin is not diminished. There are no feelings of contraction, stretching, or tightness of the skin. There are no subjective symptoms whatsoever.

Having thus given a general description of the disease, we may examine more closely the several regions invaded, pointing out any peculiarities that may be worthy of mention. The hair of the head is dry, but not remarkably so. The scalp is free of seborrhœa. It is well covered with disseminated freckles, which give it a dirty appearance, and which look like small collections of dirty sebaceous matter. They occupy the whole of the scalp, but are not so profuse as upon the face and other regions. They are in greatest numbers over the vertex and on the parietal regions. About the occiput they are thinly scattered. They differ in no way from those on the face, to be presently described. Here and there occur small telangiectases, some of which are typically developed, while others are in a shrunken state, and apparently passing into atrophic spots. Well-marked, complete atrophic lesions exist only here and there; several are observed in the median line where the hair is parted.

The face is well sown with freckles of all colours, but it is by no means so extensively invaded as the neck, chest, back, or arms; nor are the lesions here as large as on the regions just specified. They are least numerous upon the nose and upper part of the cheeks, where exist distinct, mostly superficial, atrophic marks, varying in size from a large pin-head to a split-pea. These lesions are the most pronounced of any of the atrophic ones, and are about as conspicuous as small variola scars, which indeed they resemble in size and in form, as well as in the amount of cicatricial tissue. There are about a dozen of them. They show no regularity of

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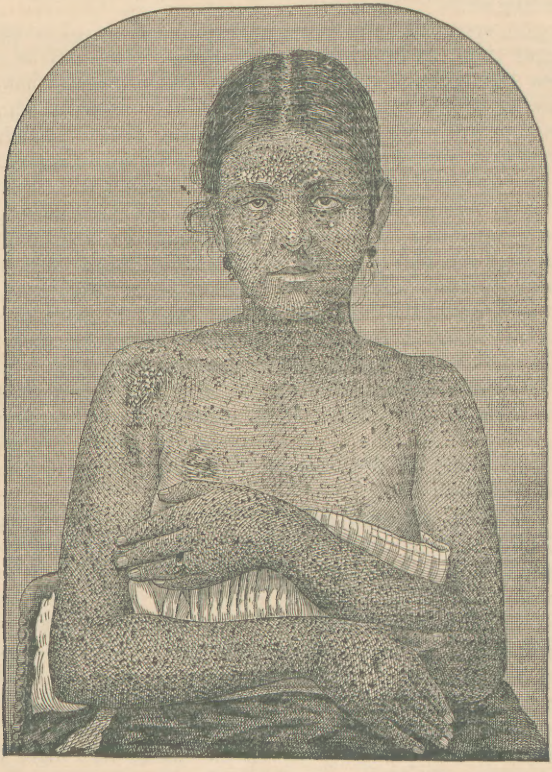
arrangement. The history of these lesions, it will be remembered, is not perfectly clear, but there is every reason to believe that they are the result of the disease under consideration, and not of the accidental eruption referred to. The telangiectases on the face are few, small, and ill-defined, and occur for the most part on the upper half of the cheeks. They number about a dozen, but only three or four are of any size, the rest being quite small and indistinct in outline. One of the largest is situated on the right temporal region. It is raised, and is about the size and shape of a caraway seed, and of a bright red colour.



The neck, back, and chest are thickly dotted with pigmentary lesions, some of which are remarkably large. Over the back they are found as low down as the lumbar vertebræ, but below the line of the scapulæ they are only thinly scattered, small, and faint in outline and in colour. Over the chest they are numerous, but are no different in character from those on the face or back. They fade away over the upper portion of the mammæ, none or very few existing below the line of the nipples. The lower portion of the thorax and the abdomen are entirely free, the skin being normal and quite fair. The telangiectases are more numerous on the neck than on the face, appearing for the most part as small pin-head sized, irregularly-shaped flat lesions, some of them evidently undergoing atrophic change. They are especially noticeable about the clavicles. Atrophic

marks exist here also, but they are small. Upon the back of the neck and over the back both the telangiectases and atrophic lesions are very insignificant.

The shoulders, arms, forearms, and backs of hands and fingers are thickly studded with freckles, being present almost in sheet, and give a decidedly variegated, checkered look to the skin. The extensor surfaces are especially attacked. The regions of the axillæ and the upper half of the arms are comparatively free. The extensor surfaces of the forearms, backs of hands, and fingers are particularly well marked with freckles of large size, and mostly dark in colour. Some are as large as large split-peas; these stand out quite conspicuously, and are noted to be situated at irregular intervals one from another of from one to two inches. Upon the backs of the fingers they extend as far down as the nails. The flexor surfaces of the fingers and the palms are entirely free. The telangiectases are rare over the arms and forearms, but are commoner upon the backs of the hands, where they are small, flat, chiefly of a linear form, and indistinctly defined. The atrophic marks, however, are numerous, and are better defined here than elsewhere, occurring here and there as irregularly shaped, split-pea sized areas of thinned, smooth, whitish skin. Taken between the fingers the skin can be felt to be thinned, but not in a striking degree. The atrophy is of the most superficial character, involving plainly only the uppermost strata of the skin. The condition is not unlike the senile atrophy often met with in patches on the backs of the hands of old men who have been exposed to an outdoor life. The patient is not aware how long these atrophic lesions have existed, but she remembers that formerly the backs of the hands were much more freckled than now, and she states that the whitish, cicatricial areas have developed very gradually through a period of years. She does not recall ever having observed telangiectases here, but inasmuch as she is far from being a close observer they may nevertheless have preceded the present lesions, which, I think, was without doubt the case.

The buttocks and thighs are free of disease, but the lesions again show themselves sparsely about the knees. The legs, especially the extensor surfaces, from the knees to the ankles, and the backs of the feet, are invaded with pigmentary lesions only. No telangiectases nor atrophic spots are to be found. The freckles are numerous, but are much smaller, less distinct in outline, and lighter in colour than those on the arms. The backs of the toes and the soles are not involved.

Remarks.—We have in the disease under consideration unquestionably a very rare manifestation. The case described is the first example of the affection that I have met with in Philadelphia. Some few months since, through the courtesy of Dr. R. W. Taylor, of New York, I had the opportunity of carefully examining three other cases of the same disease, residing in New York. These cases are well known to several dermatologists of New York, and, together with other cases, formed the basis of an elaborate and exhaustive article on this disease by Dr. Taylor, which was presented at the first annual meeting of the American Dermatological Association, in Sept. 1877.[1]

[1] The article has unfortunately not as yet been published, and I am therefore debarred from referring to it more in detail.

From a review of the case presented, we may note the following points of interest. In the first place, the early age at which the disease made its appearance. This statement is entirely in accord with the history of both Hebra's and Taylor's cases, the disease in every instance having appeared in early life. The extremely slow evolution of the symptoms, and the chronic course of the affection as a process, were also noted in the above cited cases. As has been already stated, the pigmentary lesions differ in no essential particular from common lentigines. The patient was indeed on several occasions treated for freckles. The lesions, however, are not only larger but are much more deeply pigmented than in common lentigo, the blackish lesions being singularly conspicuous and peculiar. The telangiectases present nothing unusual. The atrophic marks are remarkable for their superficial character, the atrophy being confined to the uppermost layers of the corium, and in all probability not extending below the papillary layer. They may be compared to the well-known maculæ atrophicæ, but they are even more superficial, and are less markedly circumscribed, and less conspicuous.

The relation one to the other of the several kinds of lesions is a most interesting topic. From the history of the case, as well as from the present appearance of the lesions, it may, I think, be considered as pretty clearly established that the following course is the usual one. The pigmentary spots are the first symptoms, which in a variable time are here and there succeeded by the development of telangiectases, occupying as a rule the whole or greater portion of the freckle. They continue for a longer or shorter period, when they begin to contract and undergo atrophy, finally disappearing, to be followed by the atrophic macules. As the telangiectases happen to be small or large, superficial or deeper-seated, will the subsequent atrophic spots be slight or extensive, superficial or deeper.

Viewed from a pathological standpoint it is difficult to know where the disease should be placed; whether among the pigment hypertrophies, with the new growths, or with the atrophies. Hebra and Kaposi kave regarded the affection as an atrophy, and have considered it under the head of the diffused idiopathic atrophies, along with senile atrophy. They have given to it the name "xeroderma," or "parchment skin," a name which, however, as will I think be admitted, is by no means appropriate to the phase of the disease represented by the case under consideration. In their work on diseases of the skin[1] they give brief notes of two cases which had fallen under their observation, and refer in a few lines to two other cases that they had seen, being the whole material from which their knowledge of the disease is drawn. These cases constitute, I believe, the only examples of the affection that have been published. In Hebra's and Kaposi's cases (both girls, eighteen and ten years of age, in both of whom the dis-

[1] New Syd. Soc. Transl., vol. iii. p. 252.

ease had existed since early childhood), the process assumed a much severer type than in my case, the atrophy being marked and extensive. Kaposi thus speaks of the atrophy in the first case, aged eighteen: The skin "was also tightly stretched, as if contracted, was pinched up into a fold with difficulty, and felt very thin. Its surface was smooth in some places, whilst in others fine epidermic lamellæ peeled off; or there were quite flat, linear furrows marked out on the epidermis, so that the surface appeared as dry as parchment, and wrinkled, whilst the skin itself was tightly stretched." Concerning the atrophy in the second case, it is stated that "the epidermis, especially on the eyelids and on the cheeks, was wrinkled and shrivelled, the upper eyelids being in consequence drawn somewhat downwards, and the lower ones drawn down and everted, the eyes, therefore, seeming from above too small, and from below incompletely covered. In the same way the oral and nasal apertures were somewhat diminished. In addition, the skin was moderately tense, and was less readily than normally pinched up into a fold, but this could always be accomplished. The subcutaneous layer of fat was not altered." It will thus be seen that the atrophy formed a most important symptom in these two cases, a symptom, however, which was by no means either extensive or striking in my case; nor was there any indication of the process assuming any proportions similar to those presented in Hebra's and Kaposi's cases. In three of Taylor's cases, those that I saw, the atrophy, while more extensive, still manifested the same general characters noted in my case. It is therefore manifest that the term xeroderma is altogether inappropriate for the stages of the disease represented by the present case. It is, moreover, unfortunate that the term xeroderma should have been selected for this new disease, inasmuch as this word has for some years been in use as signifying a mild form of ichthyosis, and is in this sense employed to-day by many writers, both abroad and in this country. While it is admitted that the atrophy of the skin is an important feature, and in some cases the gravest symptom, in the course of the disease, it is also clear from a study of the case just considered that it is not necessarily the most prominent symptom. The pathological process is, as we have seen, a complex one, including hypertrophy, atrophy, and new growth. In view of this association of several distinct processes, occurring simultaneously or consecutively, it becomes extremely difficult to place the disease, and also to name it. Until our knowledge concerning it is more complete, it will, I think, be well to postpone the adoption of any final nomenclature.

As regards treatment, I am able to say but little. Locally the pigmentary lesions may doubtless be treated with more or less success by means of the well-known remedies used against lentigo; while the telangiectases may be removed with a solution of caustic potash (℥j to ʒj), as in the case of other small telangiectases.